Calcifying fibrous tumor arising from the heart

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Calcifying fibrous tumor (CFT) is a rare benign tumor, first reported in 1988,[1] that arises usually from the soft tissues of the extremities and rarely from the pleura, mediastinum, and lung. The lesion is histologically characterized by hypocellular, hyalinized, collagenous tissue with calcifications and lymphoplasmacytic infiltrates.[1,2] Dystrophic calcifications and psammomatous bodies are specific to this entity, distinguishing it from inflammatory myofibroblastic tumor (IMT), also benign lesion, which is usually more cellular and less hyalinized. Although the number of cases is insufficient to establish the therapeutic approach, completely surgical resection appears to be the best option and recurrence is rare. We describe here a first case of CFT originating from the left auricle of the heart in a young woman.

Clinical Summary

A 16-year-old woman who was in good health without any symptoms was referred with an abnormal chest roentgenogram at routine examination. Computed tomographic scan and magnetic resonance imaging confirmed a large and heterogeneous mass with irregular border compressing the left ventricle and left pulmonary vein in the pericardium (Figure 1). A biopsy via video-assisted thoracoscopy was performed to confirm the histologic diagnosis. After division of normal-appearing pericardium, a lobulated, elastic, firm, and pale mass was found on the left ventricle, with serous pericardial effusion. The biopsy sample showed collagenous stroma with spindle cells and inflammatory cells, which supported a possible diagnosis of IMT. No cytologic abnormalities were observed in the pericardial fluid. Because of the possibility of malignant transformation and to prevent the appearance of clinical symptoms related to enlarged tumors, the tumor was resected. At operation, the heart was approached through a left lateral thoracotomy. The lobulated mass compressed the left ventricle but showed no invasive growth to the adjacent tissues. The base of the tumor was attached on the left auricle of the heart, and the remaining tumor was completely resected without cardiopulmonary bypass. The outer surface was relatively smooth and lobulated. The cut surface revealed a white, homogeneous, and firm appearance, without evidence of necrosis or hemorrhage. Histologically, the tumor was composed of collagenous tissues containing spindle cells, psammomatous calcifications, and infiltrates with lymphocytes, with no necrosis (Figure 2). No features of malignancy were seen on light microscopy. The tumor cells yielded negative immunohistochemical results for smooth muscle actin, desmin, and anaplastic lymphoma kinase (ALK). These findings were consistent with a diagnosis of CFT rather than IMT.

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Discussion

CFT is a very rare lesion, and its etiology is still unclear. Only 12 cases in the thorax—including the pleura, the mediastinum, the lung, and the chest wall—have been reported.[3] The case described here is unique in its site of origin, and to our knowledge this is the first report of CFT arising from the heart. No cardiac symptoms

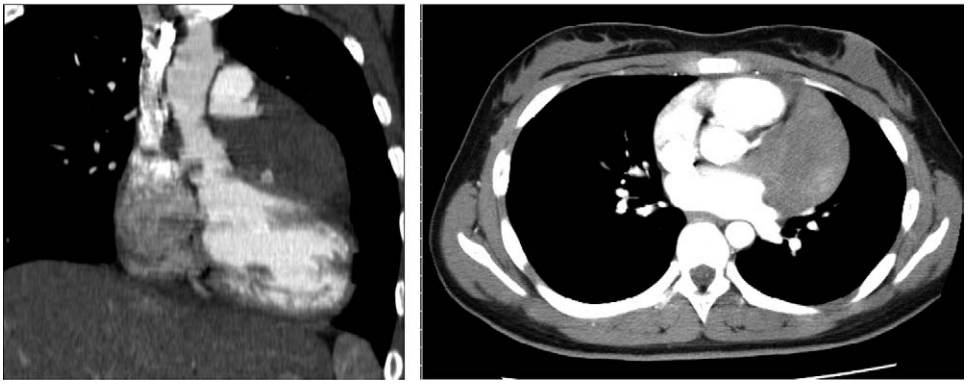

Figure 1. Computed tomographic scans showing large, heterogeneous mass compressing left ventricle in pericardium.

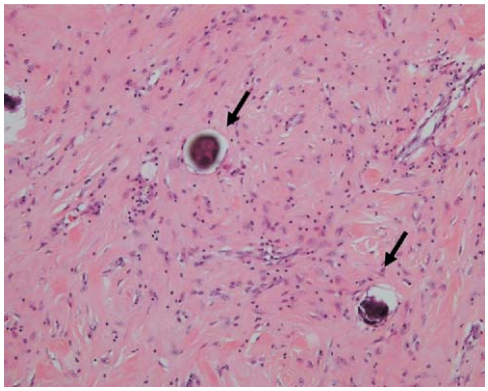

Figure 2. Tumor was composed of collagenous tissues containing spindle cells, psammomatous calcifications *(arrows)*, and infiltrates with lymphocytes.

were observed in this case, even though the mass compressed the left atrium and ventricle. Since we could not exclude the possibility of malignant tumor, an immediate histologic diagnosis was required. One should keep this entity in mind when making the differential diagnosis of tumors arising from the heart.

Because it has been postulated that CFT is possibly a late sclerosing stage of IMT,[4] histologically distinguishing CFT from IMT may be difficult. Recently, some IMTs have been shown to have chromosomal translocation involving the *ALK* gene located on chromosome 2p, as seen in anaplastic large cell lymphoma. These ALK fusion proteins are constitutively activated, are associated with aberrant expression (immunopositive for ALK), and have potentially oncogenic property.[5] There have been no genetic or molecular studies of CFT so far. Further examinations including molecular approaches should be conducted to understand the pathogenesis of CFT as well as IMT.

With the efficacy of chemotherapy and radiotherapy remaining unknown, complete resection appears to be the best option if the mass is surgically resectable. Although a few cases of local recurrence have been reported, there have been no reports to date of distant metastases. At present, there are no valuable predictive factors for the biologic and clinical behavior of CFT. Careful and close follow-up is mandatory.

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